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Complete heart block and systemic lupus erythematosus

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An 18-year-old girl with systemic lupus erythematosus developed progressive electrocardiographic abnormalities over a period of 16 years, culminating in complete heart block with Adams-Stokes attacks. A permanent ventricular pacing system was implanted successfully.

Although cardiac conduction disturbances in systemic lupus erythematosus are not uncommon, complete heart block is rare. Moffitt (1965) described a patient with this complication who required permanent pacemaker implantation. This report records the gradual development of conduction disturbances leading to complete heart block in another patient with systemic lupus erythematosus.

Case report

An 18-year-old typist presented in March 1958 with a 7-month history of recurrent attacks of joint stiffness and swelling affecting especially the small joints of the hands, but also the shoulders and knees. She had recently developed Raynaud's phenomenon. Physical examination showed peripheral polyarthritis, with slight flexion deformity of the fingers and a poor peripheral circulation. Haemoglobin was 10.6 g/dl, white cell count $6.4 \times 10^3/\mu l$, ESR 42 mm in the first hour (Westergren). Urine testing for protein was negative. Chest *x*-ray showed an increase in size of the cardiac silhouette; *x*-rays of the hands and feet were normal. An electrocardiogram was normal with a PR interval of 0.16 s and an ÂQRS of +67° (Fig.).

In August 1958 she was admitted to this hospital with increasing joint pain. On examination there was facial erythema, pyrexia (39.5°C), tachycardia (100/min regular), a pericardial friction rub, a right pleural effusion, ulnar deviation of the fingers, impaired wrist and finger movement, and an effusion in the right knee. The ESR was 80 mm in the first hour and occasional lupus erythematosus cells were seen in peripheral blood *in vitro* preparations, but not in the bone marrow aspirate. Chest *x*-ray showed a further increase in the cardiac silhouette in keeping with a pericardial effusion, while serial electrocardiograms revealed a progressive fall in voltage. Steroid therapy with prednisolone 30 mg daily resulted in rapid resolution of the pericardial effusion and an increase in electrocardiographic voltage. Prednisolone was discontinued in November 1958 but reintroduced in 1959 when the joints again deteriorated with fixed flexion deformities at the proximal interphalangeal joints. The lupus erythematosus cell test remained intermittently positive with hyperglobulinaemia (50 g/l; increased gammaglobulin on electrophoresis) and a persistently raised ESR. Because of the history, the symptoms, the malar erythema, the capillary loops visible at the finger nail folds, the raised serum globulin and ESR, the diagnosis was considered to be systemic lupus erythematosus. Since the response to prednisolone was unsatisfactory, chloroquine sulphate, 250 mg b.d. was given in addition.

Between 1959 and 1970 the lupus erythematosus cell test was occasionally positive but the differential agglutination test was positive only once (1:32) and the Wassermann reaction repeatedly negative. Barium swallow examination in 1968 was normal. *X*-rays of the hands showed ulnar deviation but no evidence of osteoporosis or bony erosions. There was tufting of the terminal phalanges. Electrocardiographic examination in 1964 showed a change in ÂQRS to −35° (Fig.).

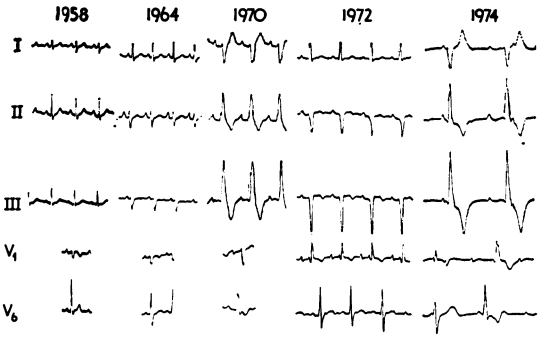

FIG.

In 1970 she presented with increasing dyspnoea and palpitations. On examination there was evidence of heart failure with an increased jugular venous pressure, cardiomegaly, and a gallop rhythm. The blood pressure was 155/105 mmHg (20.6/14.0 kPa). Chloroquine sulphate was discontinued and prednisolone 10 mg daily maintained. An electrocardiogram showed left bundle-

branch block with a PR interval increased at 0.24 s and right axis deviation (ÂQRS+112°) (Fig.). Antinuclear factor titre >1:500 (speckled pattern). Hypotensive therapy with methyldopa resulted in a satisfactory fall in blood pressure to 100/75 mmHg (13.3/10.0 kPa), and there was a symptomatic improvement with digoxin and diuretics. Methyldopa was subsequently discontinued.

In 1972 she was readmitted with a pyrexia (40.5°C) and later a loud pericardial friction rub developed, which responded to an increase in prednisolone dosage to 40 mg daily. The electrocardiogram now showed right bundle-branch block with a PR interval of 0.2 s and left axis deviation (ÂQRS −63°) (Fig.). She was discharged home taking prednisolone 20 mg daily.

Following bradycardia for three hours in March 1974 she had an Adams-Stokes attack and was admitted to hospital. An electrocardiogram showed complete heart block with QRS widening to 0.14 s (ÂQRS+120°), a ventricular rate of 44/min, and an atrial rate of 100/min (Fig.). A temporary pacemaker was inserted before a permanent pacing system (Devices 3821 RC) was implanted in the abdomen. She was discharged home in regular paced rhythm.

The antinuclear factor was positive (1 in 100, speckled pattern), DNA binding (nDNA) increased at 63 per cent (normal value 0 to 30%: Dr. G. Hughes), C_3 62 mg/100 ml (normal 104–161 mg/100 ml); creatinine phosphokinase 22 IU/l (normal range 0–60 IU/l); haemoglobin 11.8 g/dl, ESR 35 mm in the first hour; chest *x*-ray showed obvious cardiomegaly; blood urea 6.5 mmol/l; urine testing for protein negative.

Discussion

The conduction disturbances seen in systemic lupus erythematosus are rarely as extensive as in the present case. The sinoatrial and atrioventricular nodes may be involved. Bundle-branch block is rare. The histological changes have been documented by James, Rupe, and Monto (1965): Case 3 of his report describes a 48-year-old white woman who developed complete heart block and subsequently died; on microscopical examination many inflammatory cells were seen at the interface between the atrioventricular node and the central fibrous body. Two other patients reported in this series died suddenly and a fatal Adams-Stokes attack cannot be excluded from the data presented.

The clinical improvement resulting from cardiac pacing was gratifying in this patient. It is possible that with the widespread use of long-term steroid therapy in systemic lupus erythematosus other instances of complete heart block caused by this may be found.

Our thanks are due to Dr. Maxwell Telling for permission to report this case, and to Dr. G. Hughes for nDNA binding studies.

References

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